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Adrenocortical carcinoma arising from the colonic mesentery

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ABSTRACT

A 57-year-old woman presented with left-sided abdominal pain and was found to have a large retroperitoneal mass in the left upper quadrant. Surgical excision and histopathologic evaluation demonstrated an adrenocortical carcinoma arising from the colonic mesentery. Ectopic adrenocortical carcinomas are extremely rare tumors with an unknown incidence. Functioning tumors typically present with the Cushing syndrome and/or virilization, whereas nonfunctioning tumors result in abdominal pain from mass effect. They are highly aggressive tumors with an overall poor prognosis.

KEYWORDS Adrenocortical carcinoma; ectopic; mesentery

Primary adrenocortical carcinoma (ACC) is a rare malignant tumor arising from the adrenal cortex with an incidence of 1 to 2 cases per million per year.[1] Ectopic location of the tumor is even more rare, with an unknown incidence and prevalence. We present a symptomatic woman found to have an ACC arising from the colonic mesentery.

CASE DESCRIPTION

A 57-year-old woman presented to the emergency department with a 2-week history of left-sided abdominal pain. Physical examination demonstrated tenderness to palpation in the left upper quadrant. Laboratory findings were consistent with anemia of chronic disease. Computed tomography (CT) of the abdomen and pelvis showed a large retroperitoneal mass within the left hemiabdomen *(Figure 1)*. Surgical excision showed a large mobile mass in the left upper quadrant attached to the colonic mesocolon and distinct from adjacent solid organs. Gross pathology demonstrated a 459 g, 11 cm lobulated yellow mass with a red rubbery outer surface and diffuse hemorrhage. Microscopy showed a hypercellular tumor composed of round to ovoid cells with focally microcystic patterns *(Figure 2)*. The tumor cells showed uniform nuclei with vesicular chromatin and scant pale to amphophilic cytoplasm. Marked mitoses (up to 24 per 50 high-power fields), patchy tumor necrosis, focal capsular invasion, and vascular invasion were present with an overall Modified Weiss Criteria score of 5. Immunohistochemistry was positive for Melan-A, inhibin, synaptophysin, Cam 5.2, calretinin, and S100 and negative for chromogranin. Histopathologic findings were overall consistent with ACC. Magnetic resonance imaging (MRI) of the abdomen performed 6 months postoperatively showed new lesions in the liver concerning for metastases *(Figure 3)*. Subsequent partial hepatic lobectomy was performed with histopathologic findings consistent with metastatic ACC. The patient is currently under close monitoring with serial imaging.

DISCUSSION

Primary ACC is a malignant tumor arising from the adrenal gland, specifically the adrenal cortex. The two primary components of the adrenal gland, the adrenal cortex and adrenal medulla, are formed by separate embryologic origins. The cortex is derived from the urogenital ridge and celomic epithelium, while the medulla is derived from neural crest cells.[2,3] The urogenital ridge is a structure within the embryologic mesoderm that also gives rise to the gonads, kidneys, and reproductive tract while the celomic epithelium produces the lining of the abdominal organs and surface of the body wall.[3,4] This embryologic phenomenon is thought to explain the observation of ectopic adrenocortical tissue. However, these cases are rarely observed in adults because ectopic adrenal tissue typically atrophies during infancy.[4] Reported sites of

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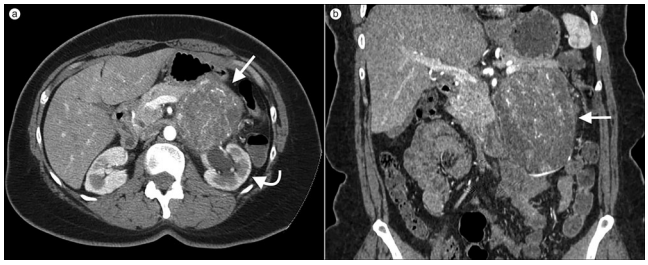

Figure 1. CT of the abdomen and pelvis with intravenous contrast in **(a)** axial and **(b)** coronal planes demonstrates a large inhomogeneous hypervascular mass in the left abdomen (straight arrow). The mass results in compression of the left ureter with moderate left hydronephrosis (curved arrow).

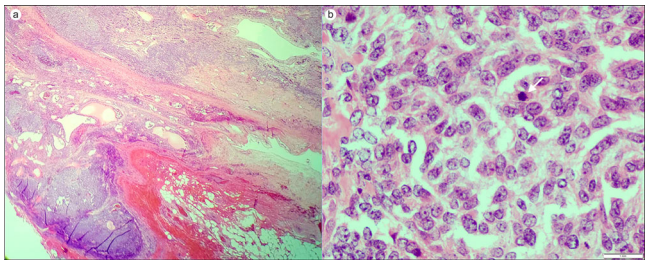

Figure 2. Microscopic images of adrenocortical carcinoma. **(a)** Low-power view shows tumor with vascular invasion. **(b)** High-power view demonstrates round to ovoid cells with a focally microcystic pattern and several mitotic figures (arrow).

ectopy include the testes, ovaries, kidneys, bowel, pancreas, liver, lungs, and mesentery.[1] To our knowledge, fewer than five cases of ectopic ACCs have been reported to arise from the mesentery, which was the case in our patient.[5–7]

Clinical presentation depends on whether the tumors are functioning or nonfunctioning. Patients with functioning tumors typically present with symptoms of the Cushing syndrome and/or virilization.[8] Many nonfunctioning tumors are detected incidentally on radiographic studies performed for another reason. Imaging generally shows an inhomogeneous hypervascular tumor with irregular margins with or without necrosis and hemorrhage.[9,10] Biopsy and subsequent histopathologic evaluation are required for diagnosis. Immunohistochemistry findings of positive alpha-inhibin, Melanin-A, and SF-1 can confirm the primary adrenal origin.[11] The Modified Weiss Criteria use histopathologic characteristics to differentiate an adrenal adenoma from carcinoma. These characteristics include a mitotic rate >5 per 50 high-power fields, clear cytoplasm compromising ≤25% of the tumor, abnormal mitoses, necrosis, and capsular invasion, with each criterion receiving a score of 0 when absent and 1 when present. Using the formula 2× mitotic rate criterion + 2× clear cytoplasm criterion + abnormal mitoses + necrosis + capsular invasion, a score of 3 or more suggests malignancy.[12]

Complete resection of the tumor is the only potentially curative treatment.[11] For patients with a high risk of recurrence, unresectable tumors, or metastasis, adjuvant chemotherapy and radiation may be considered. Mitotane, an adrenocorticolytic drug, has shown efficacy in patients with ACC.[13] Overall prognosis tends to be poor and is determined by disease stage and completeness of resection. The 5-year overall survival is 66%, 58%, 24%, and 0% for stage I, II, III, and IV disease, respectively.[14]

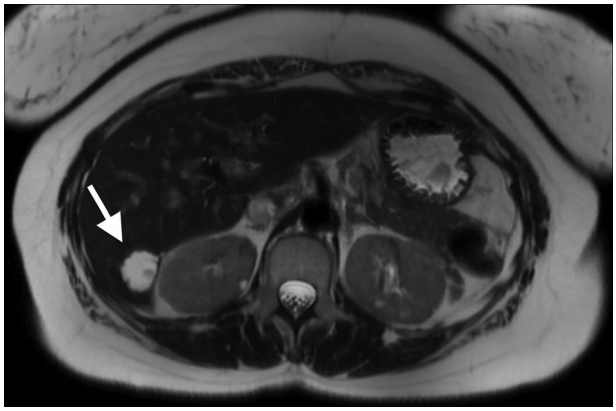

Figure 3. MRI of the abdomen in axial plane shows a lesion in the right hepatic lobe concerning for metastasis (arrow).

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